General

Guideline Title

2014 UK national guideline on the management of vulval conditions.

Bibliographic Source(s)

Clinical Effectiveness Group. 2014 UK national guideline on the management of vulval conditions. London (UK): British Association for Sexual Health and HIV (BASHH); 2014 Feb. 22 p. [60 references]

Guideline Status

This is the current release of the guideline.

This guideline updates a previous version: Clinical Effectiveness Group. 2007 UK national guideline on the management of vulval conditions. London (UK): British Association for Sexual Health and HIV (BASHH); 2007. 16 p. [44 references]

Recommendations

Major Recommendations

Levels of evidence (I-IV) and grades of recommendation (A-C) are defined at the end of the "Major Recommendations" field.

Management

General Advice for Delivery of Vulval Care

Vulval conditions may present to genitourinary medicine physicians, dermatologists and gynaecologists and treatment modalities also span across this spectrum. Care of patients with vulval conditions is therefore best managed by a multidisciplinary approach. This includes clear working arrangements between disciplines or access to a specialist multidisciplinary vulval service. The service should also have access to clinicopathological discussion and review of histology results.

General Advice for All Vulval Conditions

- Avoid contact with soap, shampoo and bubble bath. Simple emollients can be used as a soap substitute and general moisturizer.
- Avoid tight fitting garments which may irritate the area
- Avoid use of spermicidally lubricated condoms
- Patients should be given a detailed explanation of their condition with particular emphasis on any long-term implications for the health of themselves and their partner(s). This should be reinforced by giving them clear and accurate written information. (The British Society for the Study of Vulval Disease [BSSVD] Web site provides some patient information, including British Association of

Dermatology [BAD] leaflets , International Society for the Study of Vulvovaginal Disease [ISSVD], Vulval Pain Society and NZ Dermnet)
- The patient's general practitioner (GP) should be informed
- Sexually transmitted infection (STI) screening should be considered in all patients and vulvovaginal candidiasis excluded if the patient presents with vulval itch
- All patients should be assessed for sexual dysfunction

Sexual Partners

Partner tracing is not required unless screening detects an STI.

Vulval Lichen Sclerosus

These guidelines relate to the management of lichen sclerosus in adult females. Additional guidelines have been produced by the Royal College of Obstetricians and Gynaecologists and the British Association of Dermatologists.

Clinical Features

Symptoms

- Itch
- Soreness
- Dyspareunia if introital narrowing
- Urinary symptoms
- Other symptoms, e.g., constipation, can occur if there is perianal involvement
- Can be asymptomatic, but this is rare

Signs

- Pale, white atrophic areas affecting the vulva
- Purpura (ecchymosis) is common
- Fissuring
- Erosions, but blistering is very rare
- Hyperkeratosis can occur
- Changes may be localised or in a 'figure of eight' distribution including the perianal area
- Loss of architecture may be manifest as loss of the labia minora and/or midline fusion. The clitoral hood may be sealed over the clitoris so that it is buried.

Complications

- Development of squamous cell carcinoma (SCC; actual risk <5%)
- Development of clitoral pseudo cyst
- Sexual dysfunction
- Dysaesthesia

Diagnosis

- Characteristic clinical appearance
- Histology of vulval biopsy: thinned epidermis with sub-epidermal hyalinization and deeper inflammatory infiltrate. In early disease histology can be difficult.

Management

Further Investigation

- Biopsy: is mandatory if the diagnosis is uncertain, there are atypical features or coexistent vulval intraepithelial neoplasia (VIN)/SCC is suspected
- Investigation for autoimmune disease if clinically indicated, especially thyroid dysfunction (i.e., thyroxine [T4] and thyroid-stimulating hormone [TSH]) as it is often asymptomatic and has been found be associated (IV, C)
- Skin swab: only useful to exclude co-existing infection if there are symptoms or signs suggestive of this

- Patch testing: rarely required and only if secondary medicament allergy suspected. The advice of a dermatologist should be sought

Treatment

General Advice

Patients should be informed about the condition and given written information. Patients should be made aware of the small risk of neoplastic change. They should be advised to contact the doctor if they notice a change in appearance or texture (e.g., lump or hardening of skin), or if there is a major change in symptoms.

Recommended Regimen

- Ultra-potent topical steroids, e.g., clobetasol propionate (I, A). Various regimens are used, one of the most common being daily use for one month, alternate days for one month, twice weekly for one month with review at 3 months. It can then be used as needed depending on symptoms. There is no evidence on the optimal regimen.
- 30 gm of an ultra-potent steroid should last at least 3 months
- Ointment bases are much better to use on the ano-genital skin because of the reduced need for preservatives in an ointment base, and hence less risk of a secondary contact allergy

Alternative Regimens

An ultra-potent topical steroid with antibacterial and antifungal, e.g., Dermovate NN® or generic equivalent (clobetasol with neomycin and nystatin) or an alternative preparation that combats secondary infection (such as Fucibet® [betamethasone with fusidic acid] cream) may be appropriate if secondary infection is a concern. These should only be used for a short period of time to clear infection (IV, C).

Research Findings and Unlicensed Treatments

- Topical calcineurin inhibitors. This is not a licensed indication and long-term safety and efficacy is not established. Tacrolimus 0.1% has been shown to be effective when used for 16 to 24 weeks (IIb, B). This study, which included males and females and genital and extra genital lichen sclerosus, showed that 77% of evaluable patients responded to treatment with 43% showing a complete response (absence of symptoms and skin findings excepting induration and atrophy) at 24 weeks. The follow-up period was 18 months and whilst no patient was shown to have skin malignancy or dysplastic change the long-term risks need to be studied in view of concerns about the possibility of topical immunosuppression increasing susceptibility of malignancy. A study of the related agent, pimecrolimus, showed that 42% of patients were in 'complete remission' after 6 months application (IIb, B). Local irritancy was the most common side effect with both tacrolimus and pimecrolimus but usually improved after the initial period of use. Cases of malignancy have been reported and it is recommended that these agents should not be used first line.
- Oral retinoids, e.g., acitretin – these may be effective in severe recalcitrant disease (Ib, A) but should only be given by a dermatologist, experienced in the use of these agents. They are severely teratogenic and pregnancy must be avoided for 2 years after finishing treatment.
- Ultraviolet A1 (UVA1) phototherapy has been reported as successful in a small number of cases (III, B)

Pregnancy and Breast-Feeding

- Topical steroids are safe to use while pregnant or breast-feeding
- Topical calcineurin inhibitors are contraindicated whilst pregnant or breast-feeding
- Retinoids are absolutely contraindicated during pregnancy and for at least 2 years before. They should be used with caution in females of child-bearing age.

Onward Referral Criteria

- Those with active disease which has not responded adequately to treatment should be referred to a specialised vulval clinic
- Any patient who develops differentiated or undifferentiated VIN or an SCC on a background of lichen sclerosus should be seen and follow-up in a specialised vulval clinic. Surgery – should only be used for the treatment of coexistent VIN/SCC or fusion. Disease tends to recur around the scar (III, B).

Follow-Up

- After 3 months to assess response to treatment
- Stable disease should be reviewed annually and this can be done by the GP in those with well controlled disease. This must be communicated to the patient and GP by the clinic.

- Patients should be informed that if they notice the development of a lump, sore area, change in symptoms or change in appearance they should prompt medical review

Vulval Lichen Planus

Clinical Features

Symptoms

- Itch/irritation
- Soreness
- Dyspareunia
- Urinary symptoms
- Vaginal discharge
- Can be asymptomatic

Signs

The anogenital lesions of lichen planus may be divided into three main groups according to their clinical presentation:

1. Classical: typical papules will be found on the keratinised anogenital skin, with or without striae on the inner aspect of the vulva. Hyperpigmentation frequently follows their resolution, particularly those with dark skin. This type of lichen planus may be asymptomatic. Vulval lesions were found in 19 out of 37 women with cutaneous lichen planus, with four of the 19 having had no symptoms.
2. Hypertrophic: these lesions are relatively rare and can be difficult to diagnose. They particularly affect the perineum and perianal area, presenting as thickened warty plaques which may become ulcerated, infected and painful. Because of these features, they can mimic malignancy. They do not appear to be accompanied by vaginal lesions.
3. Erosive: the most common subtype to cause vulval symptoms. The mucosal surfaces are eroded. At the edges of the erosions the epithelium is mauve and a pale network (Wickham's striae) is sometimes seen. The vaginal lesions in erosive lichen planus are important to recognise early and start treatment as they can lead to scarring and complete stenosis. The lesions consist of friable telangiectasia with patchy erythema which are responsible for the common symptoms of postcoital bleeding, dyspareunia and a variable discharge which is often serosanguinous. As erosions heal synechiae and scarring can develop. This type is also seen in the oral mucosa although synechiae are uncommon. The term vulvo-vaginal gingival syndrome is used when erosive disease occurs in these three sites. The presenting symptom is usually of pain and soreness.

Complications

- Scarring, including vaginal synechiae
- Development of SCC. In one study the incidence was as high as 3%.

Diagnosis

- Characteristic clinical appearance. Involvement of the vagina excludes lichen sclerosus. Skin changes elsewhere can be helpful but overlap between lichen planus and lichen sclerosus is described. Immunobullous disorders such as pemphigus can look clinically similar to erosive lichen planus.
- Histology of vulval biopsy: irregular saw-toothed acanthosis, increased granular layer and basal cell liquefaction. Band-like dermal infiltrate mainly lymphocytic.

Management

Further Investigation

- Biopsy: is a necessity if the diagnosis is uncertain or coexistent VIN/SCC is suspected. Direct immunofluorescence should be performed if an immunobullous disease is considered in the differential diagnosis. Only 25% are classic on biopsy and clinicopathological discussion is important.
- Investigation for autoimmune disease especially of the thyroid (i.e., T4 and TSH if there is any suspicion of abnormality) (IV, C)
- Skin swab: to exclude secondary infection especially of excoriated lesions
- Patch testing: if secondary medicament allergy suspected
- Whilst a link with hepatitis C and sometimes B has been noted in some countries there is no evidence of increased incidence in the United Kingdom (UK) and routine screening is not thought necessary

Treatment

General Advice

Patients should be informed about the condition and given written information. Patients should be made aware of the small risk of neoplastic change. They should be advised to contact the doctor if they notice a change in appearance or texture (e.g., lump or hardening of skin).

Recommended Regimen

- Ultra-potent topical steroids e.g., clobetasol propionate (Ib, B). In a study of 114 patients in a vulval clinic, 89 used ultra-potent topical steroids as first line treatment of whom 75% improved and 54% were symptom free. However in only 9% was there resolution of signs of inflammation. There is no evidence on the optimal regimen.
- Maintenance treatment may be required and can either be with weaker steroid preparations or less frequent use of potent steroids
- Vaginal corticosteroids: Delivery of corticosteroids to the vagina is not easy. A proprietary preparation containing hydrocortisone (colifoam), introduced with an applicator, is useful. Prednisolone suppositories may be used in more severe cases (IV, C).

Alternative Regimens

An ultra-potent topical steroid with antibacterial and antifungal, e.g., Dermovate NN® or generic equivalent (clobetasol with neomycin and nystatin) or an alternative preparation that combats secondary infection (such as Fucibet® cream) may be appropriate if secondary infection is a concern. These should only be used for a short period of time to clear infection (IV, C).

Pregnancy and Breast-Feeding

- Topical steroids are safe to use while pregnant or breast-feeding
- Topical calcineurin inhibitors are contra-indicated whilst pregnant or breast-feeding
- Retinoids are absolutely contraindicated during pregnancy and for at least 2 years before. They should be used with caution in females of child-bearing age.

Onward Referral Criteria

Referral to a multidisciplinary vulval clinic is recommended for erosive disease and any recalcitrant cases, or those in whom systemic therapy is considered.

Systemic treatments: There is no consensus and little evidence base for the use of systemic agents. In the vulvovaginal–gingival syndrome there is general agreement that azathioprine, dapsone, griseofulvin, chloroquine and minocycline, all tried empirically, are of little or no benefit.

- Oral ciclosporin may be considered
- Retinoids can be very helpful in hypertrophic cases
- Oral steroids are used, for example prednisolone 40 mg/day, tapered off over a few weeks; courses can be repeated as necessary for severe flares
- The new biological agents have been used in oral and cutaneous disease. Basiliximab has been found to be effective but its use has not been evaluated in vulval disease. All these potentially toxic therapies need careful monitoring and are best supervised by a dermatologist in the context of a specialised clinic (IV, C).

Follow-Up

- At 2 to 3 months to assess response to treatment
- Active disease should be assessed as clinically required. Erosive lichen planus needs long term specialised follow-up (IV, C).
- Stable disease should be reviewed annually except in well-counselled patients who control their symptoms well. If review is by the GP this should be communicated to the patient and GP by the clinic.
- Patients should be informed that if they notice the development of a lump or change in appearance they should seek medical advice urgently.

Vulval Eczema

Clinical Features

Symptoms

- Vulval itch
- Soreness

Signs

- Erythema
- Lichenification and excoriation
- Fissuring

Complications

Secondary infection

Diagnosis

- Clinical presentation (as above)
- General examination of the skin to look for other signs of dermatitis

Management

Further Investigation

- Patch testing – standard battery and medicaments (III, B)
- Biopsy (IV, C) – only if atypical features or failure to respond to treatment

Treatment

Recommended Regimens

- Avoidance of precipitating factor (IV, C)
- Use of emollient soap substitute (aqueous cream should not be applied as a moisturiser due to the risk of irritant effects; a liquid paraffin-based preparation can be a suitable alternative)
- Topical corticosteroid – the choice of preparation will depend on severity, 1% hydrocortisone ointment in milder cases, or betamethasone valerate 0.025% or clobetasol propionate 0.05% for limited periods if severe or lichenified. A combined preparation containing antifungal and/or antibiotic may be required if secondary infection suspected. Apply once daily (IV, C).

Follow-Up

- As clinically indicated
- Long-term follow-up and psychological support may be needed

Lichen Simplex

Clinical Features

Symptoms

- Vulval itch
- Soreness

Signs

- Lichenification, i.e., thickened, slightly scaly, pale or earthy-coloured skin with accentuated markings, maybe more marked on the side opposite the dominant hand
- Erosions and fissuring
- Excoriations as a result of scratching may be seen
- The pubic hair is often lost in the area of scratching

Complications

Secondary infection

Diagnosis

- Clinical presentation (as above). Psoriasis of the vulva is usually less itchy and lesions are bright red, often glazed and well demarcated and frequently involves natal cleft.
- History including mental state examination where indicated
- General examination of the skin to look for other signs of psoriasis or lichen simplex elsewhere

Management

Further Investigation

- Screening for infection (e.g., *Staphylococcus aureus, Candida albicans*)
- Dermatological referral for consideration of patch testing – standard battery and medicaments (III, B)
- Ferritin (III, B)
- Biopsy (IV, C)

Treatment

Recommended Regimens

- Avoidance of precipitating factor (IV, C)
- Use of emollient soap (some people may have a reaction to Aqueous cream when it is used as an emollient. For this reason it is recommended only as a soap substitute and not an emollient).
- Topical corticosteroid – potent topical steroids are required when treating lichenified areas, e.g., betamethasone or clobetasol for limited periods. A combined preparation containing antifungal and/or antibiotic may be required if secondary infection suspected. Apply once or twice daily (IV, C).
- A mildly anxiolytic antihistamine such as hydroxyzine or doxepin at night is helpful
- The symptoms of pruritus often respond fairly quickly to a topical steroid but, unless the lichenification resolves, the itch–scratch cycle will remain and the symptoms will recur. A graduated reduction in the frequency of application of the topical steroid is helpful, over about 3 to 4 months.
- Cognitive behavioural therapy may be helpful if there are co-existing mental health issues

Follow-Up

- Mild disease - as clinically required
- Severe disease (i.e., when using potent topical steroids) – 1 month then as required

Vulval Psoriasis

Clinical Features

Symptoms

- Vulval itch
- Soreness
- Burning sensation

Signs

- Well demarcated brightly erythematous plaques
- Often symmetrical
- Frequently affects natal cleft
- Usually lacks scaling due to maceration
- Fissuring

Complications

May be worsened due to Koebner effect by irritation from urine, tight-fitting clothes or sexual intercourse.

Diagnosis

- Clinical presentation (as above)

- General examination of the skin and nails to look for other signs of psoriasis

Management

Further Investigation

Skin punch biopsy if the diagnosis is in doubt

Treatment

Recommended Regimens

- Avoidance of irritating factors
- Use of emollient soap substitute
- Topical corticosteroid – weak to moderate steroids are preferred but if insufficient to induce a response then intensive short term potent steroid such as clobetasol propionate 0.05% may be used. A combined preparation containing antifungal and/or antibiotic may be required if secondary infection suspected (e.g., Trimovate®) (IV, C).
- Coal-tar preparations – may be used alone or combined or alternated with topical steroids. However, these preparations can cause irritation and folliculitis (IV, C).
- Vitamin D analogues such as tacalcitol – alone or in combination with corticosteroid; however, their usefulness may be limited by causing irritation (IV, C)

Onward Referral Criteria

- Referral to a multidisciplinary vulval clinic is recommended for unresponsive or recalcitrant cases, or those in whom systemic therapy is considered.
- Systemic treatments: if required for severe and extensive psoriasis may help genital lesions but not recommended for isolated genital psoriasis.

Follow-Up

- Mild disease - as clinically required
- Severe disease – (i.e., when using potent topical steroids) 1 month then as required

Other Vulval Dermatoses

Many other skin conditions can affect the vulva. Where the diagnosis is not obvious patients should be referred to a combined vulval clinic or to a dermatologist.

Vulval Intraepithelial Neoplasia (VIN)

Clinical Features

Symptoms

- Lumps
- Burning and itch/irritation
- Asymptomatic
- Pain

Signs

- Clinical appearance is very variable
- Raised white, erythematous or pigmented lesions occur and these may be warty, moist or eroded (pigmented lesions were previously known as Bowenoid papulosis)
- Multifocal lesions are common

Complications

- Development of SCC has been reported in between 9% and 18.5% of women
- Recurrence is common and progression to cancer can occur following previous treatment

- Psychosexual consequences have also been described (especially following surgical treatment)

Diagnosis

Biopsy – histology shows loss of organisation of squamous epithelium with a variable degree of cytological atypia which is graded as undifferentiated or differentiated and by depth. Multiple biopsies may be required as there is a risk of missing invasive disease.

Management

Further Investigation

- Ensure that cervical cytology remains up to date - there is an association with cervical intraepithelial neoplasia (CIN) (this is probably only applicable to those due to human papillomavirus [HPV]) (IV, C).
- All patients with VIN should be referred for up-to-date colposcopy to exclude CIN and VIN. If there are any perianal lesions, referral for anoscopy is recommended (IV, C).

Treatment

Most studies and research relate to full thickness VIN. Multifocal lesions can be treated in the same manner as single lesions, but may have a higher recurrence rate.

Recommended Regimen

- Local excision – this is the treatment of choice for small well circumscribed lesions as it has the lowest rate of recurrence on follow up (III, B)
- Imiquimod cream 5% – partial and complete clinical and histological regression has been shown in small studies but treatment limited by side effects. Only short term follow-up data is available. This is an unlicensed indication (Ib, A).
- Vulvectomy – this has been effective but recurrence may occur and function and cosmesis will be impaired (IV, C)

Alternative Regimens

1. Local destruction – a variety of techniques have been evaluated including carbon dioxide laser and ultrasonic surgical aspiration, photodynamic therapy, cryotherapy, laser (IIa, B). There are anecdotal reports of treatment with diathermy. Involvement of skin appendages can occur and recurrence may ensue if the appropriate depth of treatment is not achieved. The recurrence rates at follow-up tend to be higher than for excision, but cosmesis is usually good.
2. 5-fluorouracil cream – may lead to resolution of some lesions but results are variable and side effects are common. No consensus on usefulness or regimen. This is an unlicensed indication (IV, C).
3. Supervision – some lesions will spontaneously regress. This may be the best policy for partial thickness VIN. However there is a risk of progression and patients should be made aware of this (IV, C).

Pregnancy and Breast-feeding

Imiquimod and 5-fluorouracil creams are not licensed in pregnancy.

Onward Referral

Cases of VIN should be assessed in a multidisciplinary vulval clinic, or have input from gynaecology regarding assessment for surgical excision.

Follow-Up

Close follow-up is mandatory. Although resolution may occur VIN III particularly has a significant rate of progression (6.5% in one study).

Vulval Pain

ISSVD defines vulvodynia as 'vulvar discomfort, most often described as burning pain, occurring in the absence of relevant visible findings or a specific, clinically identifiable, neurologic disorder'.

Vulvodynia is categorised, by the ISSVD, as generalised or localised; provoked, unprovoked or a mixture of the two.

Localised Provoked Vulvodynia (Vestibulodynia)

Clinical Features

Symptoms

Vulval pain – frequently felt at the introitus at penetration during sexual intercourse or on insertion of tampons. There is usually a long history.

Signs

- Focal tenderness elicited by gentle application of a cotton wool tip bud at the introitus or around the clitoris
- There are no signs of an acute inflammatory process

Complications

- Sexual dysfunction
- Psychological morbidity

Diagnosis

Clinical diagnosis made on history and examination

Management

Further Investigation

After exclusion of other treatable causes no further investigation is required.

Treatment

The BSSVD recommends a multidisciplinary approach to patient care and that combining treatments can be helpful in dealing with different aspects of vulval pain.

<u>Recommended Regimens</u>

- Avoidance of irritating factors
- Use of emollient soap substitute
- Topical local anaesthetics, e.g., 5% lidocaine ointment or 2% lidocaine gel should be used with caution as irritation may be caused. The application should be made 15 to 20 minutes prior to penetrative sex and washing off the lidocaine just before sex or the use of condom by the partner can reduce the risk of transfer resulting in penile numbness. Oral contact should be avoided (IV, C).
- Physical therapies:
    - Pelvic floor muscle biofeedback (III, B)
    - Vaginal transcutaneous electrical nerve stimulation (TENS) (Ib, A)
    - Vaginal trainers (III, B)
- Cognitive behavior therapy (III, C)

<u>Alternative Regimens</u>

- Pain modifiers – the benefit of drugs such as tricyclic antidepressants, gabapentin and pregabalin is not clear. Amitriptyline gradually titrated from 10 mg up to 100 mg according to response and side effects may be beneficial in some women (IV, C).
- Surgery – Modified vestibulectomy may be considered in cases where other measures have been unsuccessful. Patients who have responded to topical lidocaine prior to sex have a better outcome (III, B).

Follow-Up

- As clinically required
- Long-term follow-up and psychological support may be needed

<u>Unprovoked Vulvodynia</u>

Clinical Features

Symptoms

- Pain that is longstanding and unexplained
- May be associated with urinary symptoms such as interstitial cystitis

Signs

The vulva appears normal.

Complications

- Sexual dysfunction
- Psychological morbidity

Diagnosis

Clinical diagnosis made on history and examination having excluded other causes.

Management

Further Investigation

After exclusion of other treatable causes no further investigation is required.

Treatment

BSSVD recommends a multidisciplinary approach to patient care and that combining treatments can be helpful in dealing with different aspects of vulval pain.

Treatment resistant unprovoked vulvodynia may require referral to a pain clinic.

Recommended Regimens

- Use of emollient soap substitute
- Pain modifiers – tricyclic antidepressants are well established in chronic pain management. Few studies have specifically examined the effect in vulvodynia however amitriptyline is frequently first line treatment; dosage should be increased by small increments starting at 10 mg up to 100 mg daily according to the patient's response (III, B).
  Note: a recent randomised study has not confirmed the beneficial effect of amitriptyline (Ib, A).
- If unresponsive or unable to tolerate the side effects, gabapentin (III, B) or pregabalin may be used (IIb, B)

Alternative Regimens

- Topical local anaesthetic, e.g., 5% lidocaine ointment or 2% lidocaine gel. A trial of local anaesthetic may be considered although irritation is a common side effect (IV, C)
- Cognitive behavioural therapy and psychotherapy (IIb, B)
- Acupuncture (IIb, C)

Follow-Up

As clinically required

Definitions:

Levels of Evidence

Level	Type of Evidence
Ia	Evidence obtained from meta-analysis of randomised controlled trials
Ib	Evidence obtained from at least one randomised controlled trial
IIa	Evidence obtained from at least one well-designed controlled study without randomisation
IIb	Evidence obtained from at least one type of well-designed quasi-experimental study
III	Evidence obtained from well-designed, non-experimental descriptive studies, such as comparative studies, correlation studies and case control studies

Level	Type of Evidence
IV	Evidence obtained from expert committee reports or opinions and/or clinical experience of respected authorities

Grades of Recommendation

Grade	Recommendation
A (Evidence levels Ia, Ib)	Requires at least one randomised controlled trial as part of the body of literature of overall good quality and consistency addressing the specific recommendation
B (Evidence levels IIa, IIb, III)	Requires availability of well-conducted clinical studies but no randomised clinical trials on the topic of recommendation
C (Evidence level IV)	Requires evidence from expert committee reports or opinions and/or clinical experience of respected authorities. Indicates absence of directly applicable studies of good quality

Clinical Algorithm(s)

None provided

Scope

Disease/Condition(s)

Non-infective vulval conditions, including:

- Vulval lichen sclerosus
- Vulval lichen planus
- Vulval eczema
- Vulval lichen simplex
- Vulval psoriasis
- Vulval intraepithelial neoplasia
- Vulval pain syndromes

Guideline Category

Counseling

Diagnosis

Evaluation

Management

Treatment

Clinical Specialty

Allergy and Immunology

Dermatology

Family Practice

Internal Medicine

Obstetrics and Gynecology

Intended Users

Advanced Practice Nurses

Nurses

Physician Assistants

Physicians

Guideline Objective(s)

To offer recommendation on the management of patients with a range of vulval disorders who may present to genitourinary medicine clinics

Target Population

Women aged 16 years or older presenting to genitourinary medicine clinics in the United Kingdom with non-infective vulval conditions

Interventions and Practices Considered

Diagnosis/Evaluation

1. History and examination including mental state
2. Sexually transmitted infection (STI) screening
3. Assessment of sexual function
4. Assessment of symptoms, signs and complications
5. Vulval biopsy and histology
6. Investigation for autoimmune disease, especially thyroid dysfunction (thyroxine [T4] and thyroid-stimulating hormone [TSH])
7. Skin swab
8. Patch testing

Treatment/Management

1. Use of a multidisciplinary approach in management
2. Counselling patients on avoiding soaps, tight fitting garments, spermicidally lubricated condoms
3. Use of emollient soap substitutes
4. Giving patients a detailed explanation of their condition with particular emphasis on any long-term implications for the health of themselves and their partners
5. Ultra-potent topical steroids (e.g., clobetasol propionate) with or without antibacterial and antifungal agents
6. Weak to moderate topical corticosteroids
7. Topical calcineurin inhibitors
8. Oral retinoids (e.g., acitretin)
9. Ultraviolet A1 (UVA1) phototherapy
10. Vaginal corticosteroids
11. Oral ciclosporin
12. Oral steroids
13. Basiliximab
14. Mildly anxiolytic antihistamine such as hydroxyzine or doxepin
15. Cognitive behavioral therapy (if mental health issues exist)
16. Acupuncture
17. Coal tar preparations
18. Vitamin D analogues such as tacalcitol

19. Surgery: local excision or vulvectomy, laser destruction, modified vestibulectomy
20. Imiquimod or 5-fluorouracil cream
21. Topical local anaesthetics (e.g., 5% lidocaine ointment or 2% lidocaine gel)
22. Pelvic floor muscle biofeedback
23. Vaginal trainers
24. Vaginal transcutaneous electrical nerve stimulation (TENS)
25. Pain modifiers (tricyclic antidepressants, gabapentin, pregabalin, amitriptyline)
26. Treatment considerations during pregnancy and breast-feeding
27. Specialist referral (if indicated)
28. Follow-up

Major Outcomes Considered

- Rate of response to treatment
- Rate of progression to dysplasia or carcinoma
- Rate of recurrence
- Complications of disease

Methodology

Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

Search Strategy

This document was produced in accordance with the guidance set out in the Clinical Effective Group's document "Framework for guideline development and assessment" (see the "Availability of Companion Documents" field). Four reference sources were used to provide a comprehensive basis for the guideline:

1. MEDLINE and EMBASE Search up to March 2012
   The search strategy comprised the following terms in the title or abstract:
   - Vulval lichen sclerosus
   - Vulval lichen planus
   - Vulval eczema
   - Vulval lichen simplex
   - Vulval psoriasis
   - Vulval intraepithelial neoplasia
   - Vulval pain syndromes/vulvodynia
2. Green-top Guideline 58 The Management of Vulval Skin Disorders. 2011 (http://www.rcog.org.uk/files/rcog-corp/GTG58Vulval22022011.pdf)
3. British Association of Dermatology Guidelines
4. Cochrane Collaboration Databases (www.cochrane.org)

Methods

Article titles and abstracts were reviewed and if relevant the full text article obtained. Priority was given to randomised controlled trial and systematic review evidence.

Number of Source Documents

Not stated

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Levels of Evidence

Level	Type of Evidence
Ia	Evidence obtained from meta-analysis of randomised controlled trials
Ib	Evidence obtained from at least one randomised controlled trial
IIa	Evidence obtained from at least one well-designed controlled study without randomisation
IIb	Evidence obtained from at least one type of well-designed quasi-experimental study
III	Evidence obtained from well-designed, non-experimental descriptive studies, such as comparative studies, correlation studies and case control studies
IV	Evidence obtained from expert committee reports or opinions and/or clinical experience of respected authorities

Methods Used to Analyze the Evidence

Systematic Review

Description of the Methods Used to Analyze the Evidence

Review of relevant research not limited to controlled trials.

Methods Used to Formulate the Recommendations

Expert Consensus

Description of Methods Used to Formulate the Recommendations

Guideline development is undertaken by a multi-disciplinary writing committee with membership determined in a transparent manner. The chair is chosen by the Clinical Effectiveness Group (CEG). The CEG lead then discusses with the chair what suggestions they might have for members from other disciplines. The additional members of the group are then invited by the CEG. Writing committee membership includes relevant professional groups (for example genitourinary medicine physicians, nurses, health advisors, pharmacists, microbiologists and other professionals from allied specialities as appropriate) and when relevant this will involve working with the appropriate British Association for Sexual Health and HIV (BASHH) Special Interest Group (SIG) and the BASHH audit group.

Patients' views and preferences are sought and considered and the process documented. This may include patient representative involvement in the writing committee, information obtained from patient interview or surveys during the writing and/or piloting process, reviewing published work on patient experiences or involving patient associations. The chair of the writing group identifies an appropriate member such as the Health Advisor to get patient feedback on the guideline. BASHH is currently developing a public panel to assist with its work and in the future this group could be approached to assist in guideline development.

Recommendations are formulated with consideration of their health benefits, side effects and risks, with evidence presented in the guideline that these issues have been addressed. Each recommendation is linked to the supporting evidence with a list of relevant references.

Consideration is given to pragmatic and organisational issues relevant to the guideline. This is sought during and may emerge from the piloting of the guideline.

The authors consider the financial cost implications of recommendations made. Where disagreement arises within the writing committee with regard to recommendations the chair attempts to resolve these (for example by a voting system or formal consensus method). The process is documented and reported to the CEG editor. When this is not possible the CEG will review the evidence.

Rating Scheme for the Strength of the Recommendations

Grading of Recommendations

Grade	Recommendation
A (Evidence levels Ia, Ib)	Requires at least one randomised controlled trial as part of the body of literature of overall good quality and consistency addressing the specific recommendation
B (Evidence levels IIa, IIb, III)	Requires availability of well-conducted clinical studies but no randomised clinical trials on the topic of recommendation
C (Evidence level IV)	Requires evidence from expert committee reports or opinions and/or clinical experience of respected authorities. Indicates absence of directly applicable studies of good quality

Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

External Peer Review

Internal Peer Review

Description of Method of Guideline Validation

This guideline has been reviewed and approved by an expert patient, and also by the British Association Sexual Health and HIV (BASHH) patient and public engagement panel.

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for selected recommendations (see the "Major Recommendations" field).

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

Appropriate management of patients with vulval conditions

Potential Harms

- Oral retinoids, e.g., acitretin – these may be effective in severe recalcitrant disease but should only be given by a dermatologist, experienced in the use of these agents. They are severely teratogenic and pregnancy must be avoided for 2 years after finishing treatment.
- Retinoids should be used with caution in females of child-bearing age
- Basiliximab has been found to be effective but its use has not been evaluated in vulval disease. All these potentially toxic therapies need careful monitoring and are best supervised by a dermatologist in the context of a specialised clinic.
- Delivery of corticosteroids to the vagina is not easy. A proprietary preparation containing hydrocortisone (colifoam), introduced with an applicator, is useful.
- Imiquimod cream 5% has shown partial and complete clinical and histological regression in small studies but treatment is limited by side effects
- Vulvectomy has been effective but recurrence may occur and function and cosmesis will be impaired
- The usefulness of vitamin D analogues such as tacalcitol, alone or in combination with corticosteroid, may be limited by causing irritation
- Topical local anaesthetics e.g., 5% lidocaine ointment or 2% lidocaine gel should be used with caution as irritation may be caused. The application should be made 15 to 20 minutes prior to penetrative sex and washing off the lidocaine just before sex or the use of condom by the partner can reduce the risk of transfer resulting in penile numbness. Oral contact should be avoided.
- With topical calcineurin inhibitors, local irritancy was the most common side effect with both tacrolimus and pimecrolimus but usually improved after the initial period of use. Cases of malignancy have been reported and it is recommended that these agents should not be used first line.

Contraindications

Contraindications

- Topical calcineurin inhibitors are contraindicated whilst pregnant or breast-feeding
- Retinoids are absolutely contraindicated during pregnancy and for at least 2 years before

Qualifying Statements

Qualifying Statements

These guidelines concentrate on a selected group of conditions, which may be managed by genitourinary physicians, either alone or in conjunction with other specialists. It is not intended as a comprehensive review of the treatment of all vulval disease.

Implementation of the Guideline

Description of Implementation Strategy

An implementation strategy was not provided.

Implementation Tools

Audit Criteria/Indicators

For information about availability, see the *Availability of Companion Documents* and *Patient Resources* fields below.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Getting Better

Living with Illness

IOM Domain

Effectiveness

Patient-centeredness

Identifying Information and Availability

Bibliographic Source(s)

Clinical Effectiveness Group. 2014 UK national guideline on the management of vulval conditions. London (UK): British Association for Sexual Health and HIV (BASHH); 2014 Feb. 22 p. [60 references]

Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2007 (revised 2014 Feb)

Guideline Developer(s)

British Association for Sexual Health and HIV - Medical Specialty Society

Source(s) of Funding

This guideline was commissioned, edited and endorsed by the British Association for Sexual Health and HIV (BASHH) Clinical Effectiveness Group (CEG) without external funding being sought or obtained.

Guideline Committee

Clinical Effectiveness Group (CEG)

Composition of Group That Authored the Guideline

Guideline Development Group: Sarah K Edwards (Consultant Genitourinary [GU] Physician); Christine Bates (Consultant GU Physician); Fiona

Lewis (Consultant Dermatologist); Cindy Sethi (Consultant GU Physician)

Lead Editor from the Clinical Effectiveness Group (CEG): Dr Deepa Grover (Consultant GU Physician)

Financial Disclosures/Conflicts of Interest

All members of the guideline writing committee completed the British Association for Sexual Health and HIV (BASHH) conflict of interest declaration detailed below at the time the guidelines final draft was submitted to the Clinical Effectiveness Group (CEG).

Guideline Endorser(s)

Royal College of Obstetricians and Gynaecologists - Medical Specialty Society

Guideline Status

This is the current release of the guideline.

This guideline updates a previous version: Clinical Effectiveness Group. 2007 UK national guideline on the management of vulval conditions. London (UK): British Association for Sexual Health and HIV (BASHH); 2007. 16 p. [44 references]

Guideline Availability

Electronic copies: Available in Portable Document Format (PDF) from the British Association of Sexual Health and HIV (BASHH) Web site .

Availability of Companion Documents

The following is available:

- Clinical Effectiveness Group. British Association for Sexual Health and HIV: framework for guideline development and assessment. London (UK): British Association for Sexual Health and HIV (BASHH); 2010. 18 p. Electronic copies: Available in Portable Document Format (PDF) from the British Association for Sexual Health and HIV (BASHH) Web site .

In addition, auditable outcomes are provided in the original guideline document .

Patient Resources

None available

NGC Status

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